## 1) THE DISEASE AND ITS EPIDEMIOLOGY

## A. Etiologic Agent

Botulism is caused by exposure to a neurotoxin produced by *Clostridium botulinum*. *C. botulinum* is an anaerobic, spore-forming bacterium. The toxin is produced as the bacteria are multiplying, and the bacteria multiply under anaerobic (no oxygen) conditions and low acid (generally pH>4). There are seven types of botulinum toxin (A–G), but human botulism is primarily caused by types A, B and E.

## **B.** Clinical Description

*C. botulinum* toxin is one of the most potent lethal substances known. In humans, botulism manifests itself in one of four clinical forms: foodborne botulism, wound botulism, infant (intestinal) botulism, and, rarely adult infectious (intestinal) botulism. The site of toxin production is different for each of the forms, but they all share the flaccid paralysis that results from exposure to botulinum toxin.

**Foodborne botulism** is a severe poisoning caused by the ingestion of pre-formed *C. botulinum* toxin. The clinical syndrome is dominated by neurologic signs and symptoms, including blurred or double vision, dysphagia, dry mouth and peripheral muscle weakness. Symmetric descending flaccid paralysis is classic for botulism, beginning with the cranial nerves. Paralysis then affects the upper extremities, the respiratory muscles and, finally the lower extremities. Patients usually require ventilatory support, which is commonly needed for 2 to 8 weeks. The clinical symptoms are similar no matter which toxin type is responsible for the illness, but type A has been associated with a higher case-fatality rate than B or E. In general, the case-fatality rate for foodborne botulism is 5-10%. Recovery may take months.

**Wound botulism** usually presents with the same clinical picture as foodborne botulism. In wound botulism, the organism multiplies in the wound and produces the toxin, which is then absorbed into the bloodstream.

**Infant (intestinal) botulism** has a distinctly different clinical presentation than wound and foodborne botulism. In infant botulism, the *C. botulinum* spores are ingested and the toxin is formed in the intestines in the absence of mature gastrointestinal flora. This disease is usually confined exclusively to infants less than one year of age. The earliest clinical sign in infant botulism is constipation, which is followed by poor feeding, decreased sucking, lethargy, listlessness, ptosis, difficulty swallowing, a weak cry, and lack of muscle tone giving rise to the term "floppy baby syndrome." In some cases, respiratory insufficiency and respiratory arrest may occur. Infant botulism presents with a wide range of severity, from mild illness to sudden death. Some studies suggest that infant botulism may be responsible for up to 5% of cases of sudden infant death syndrome (SIDS). Among hospitalized cases in the United States, the case-fatality rate is less than 1%.

**Adult infectious (intestinal) botulism** occurs as a result of toxin production in the intestines in a manner similar to infant botulism. Most people with adult infectious botulism are found to have suffered from a disruption of their natural intestinal flora due to abdominal surgery, antibiotic treatment or gastrointestinal tract abnormalities.

## C. Reservoirs

*C. botulinum* spores are ubiquitous in soils worldwide. The spores can survive indefinitely in soil under almost any environmental condition. Spores are also found in marine sediments.

#### D. Modes of Transmission

**Foodborne botulism** is acquired by ingesting pre-formed toxin. This usually occurs as a result of ingesting food that has been inadequately processed and then inadequately prepared before being eaten. The most frequent source is home-canned foods, but outbreaks have also been attributed to baked potatoes in foil, minced garlic in oil and sautéed onions held under a layer of butter. Tomato products, once thought to be a low-risk food due to a low pH, can no longer be dismissed as a potential vehicle. The toxin is destroyed by boiling.

**Wound botulism** occurs when wounds are contaminated with dirt or gravel containing botulism spores. Wound botulism has also been reported among chronic drug abusers.

**Infant (intestinal) botulism**, which is the most common form of botulism in the United States, occurs as a result of ingestion of the spore form of the bacteria, which then goes on to germinate and produce toxin in the intestines. This can happen through ingestion of food, soil or dust contaminated with botulinum spores. Honey often contains *C. botulinum* spores. Some cases of infant botulism have occurred in children living in areas of construction and earth disruption.

**Adult infectious (intestinal) botulism** occurs in a manner similar to infant botulism.

#### E. Incubation Period

The incubation period is variable, but neurologic symptoms of foodborne botulism usually appear within 12–36 hours (range: 6 hours to 8 days) after eating contaminated food. The median incubation period for wound botulism is generally longer than for foodborne botulism, usually 7 days, with a range of 4 to 14 days. In general, the shorter the incubation period the more severe the disease. The incubation period for infant botulism is unknown since it is usually not known when the spores were ingested.

## F. Period of Communicability or Infectious Period

No instances of person-to-person spread have ever been documented for botulism.

#### G. Epidemiology

Botulism occurs worldwide, as sporadic cases and as family and general outbreaks. In the US, since 1973 a median of 24 cases of foodborne botulism, 3 cases of wound botulism and 71 cases of infant botulism have been reported annually to the Centers for Disease Control and Prevention (CDC). Recently, use of black tar heroin by chronic injection drug users has led to a dramatic increase in the number of cases of wound botulism since 1994.

#### H. Bioterrorist Potential

C. botulinum toxins are considered a potential bioterrorist agent. If acquired and properly disseminated, botulinum toxin could cause a serious public health challenge in terms of ability to limit the numbers of casualties and control other repercussions from such an attack.

# 2) REPORTING CRITERIA AND LABORATORY TESTING SERVICES

## A. What to Report to the Massachusetts Department of Public Health

• **Report any suspicion of botulism** called to your attention. This could take the form of a healthcare provider or laboratory inquiring about botulism or botulism testing. Also report any suspected exposure to *C. botulinum* that may be bioterrorist in nature.

2 Botulism January 2001

*Note:* Since laboratory testing to confirm botulism is only performed at the State Laboratory Institute (SLI), the local board of health (LBOH) will not receive a report of a confirmed case of botulism without the knowledge of the Division of Epidemiology and Immunization.

*Note:* See Section 3) C below for information on how to report a case.

## **B.** Laboratory Testing Services Available

The State Laboratory Institute (SLI), Enteric Laboratory will test clinical specimens for the presence of both *C. botulinum* and botulinum toxin if epidemiologically and clinically indicated. The most reliable method for testing for toxin is the mouse neutralization test. (Due to the nature of the laboratory testing for *C. botulinum* toxin and bacteria, testing will only be done if epidemiologically and clinically indicated.) For more information call the Enterics Laboratory at (617) 983-6609 or the Division of Epidemiology & Immunization at (617) 983-6800 or (888) 658-2850.

The Food Microbiology Laboratory at the SLI will test food specimens for the presence of *C. botulinum* and/or toxin if epidemiologically indicated. For more information call the Food Microbiology Laboratory at (617) 983-6616.

## 3) DISEASE REPORTING AND CASE INVESTIGATION

## A. Purpose of Surveillance and Reporting

- To assist in the diagnosis and treatment of potential cases.
- To identify sources of public health concern (*e.g.*, a commercially distributed food product) and to stop transmission from such a source.
- To properly classify reported cases as foodborne, infant or wound botulism.
- To identify cases and clusters of human illness that may be associated with a bioterrorist event.

## B. Laboratory and Healthcare Provider Reporting Requirements

Please refer to the lists of reportable diseases (at the end of this manual's introductory section) for specific information.

*Note:* Due to the rarity and potential severity of botulism, the MDPH requests that information about any suspect or known case of botulism, or any suspected exposure that may be bioterrorist in nature, be **immediately reported** to the local board of health where diagnosed. If this is not possible, call the MDPH Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850 (weekdays), or (617) 983-6200 (emergency number for nights/weekends). Since botulism testing is only available at the SLI or the CDC, the state health department must be informed to approve appropriate testing.

## C. Local Board of Health Reporting and Follow-Up Responsibilities

#### 1. Reporting Requirements

Massachusetts Department of Public Health (MDPH) regulations (105 CMR 300.00) stipulate that each local board of health (LBOH) must report the occurrence of any case of botulism, as defined by Section 2) A above. Please refer to the Local Board of Health Reporting Timeline (at the end of this manual's introductory section) for information on prioritization and timeliness requirements of reporting and case investigation.

*Note:* The LBOH will not receive notification of confirmed cases without the knowledge of the Division of Epidemiology and Immunization since confirmatory testing is only available at the SLI and the CDC. However, the LBOH could be the initial recipient of a report of a *suspect* case.

January 2001 Botulism 3

## 2. Case Investigation

- a. The most important thing a LBOH can do upon learning of a suspect case of botulism, or any suspected exposure that may be bioterrorist in nature, is to immediately call the MDPH, any time of the day or night. Daytime phone numbers of the Division of Epidemiology and Immunization are (617) 983-6800 and (888) 658-2850. The emergency phone number for nights and weekends is (617) 983-6200.
- b. Case investigation of botulism in Massachusetts residents will be directed by the MDPH Division of Epidemiology and Immunization. If a bioterrorist event is suspected, the MDPH and other response authorities will work closely with LBOHs and provide instructions/information on how to proceed.
- c. Following immediate notification of the MDPH, the LBOH may be asked to assist in investigating any case(s) of botulism. (There is no official case report form that is required by a local board of health.) Use the following guidelines to assist you in the investigation.
- d. Determine type of botulism: Foodborne botulism is a true medical and public health emergency and should be investigated as such. Infant and wound botulism do not require the same urgency in investigation; therefore, it is essential to determine what illness is occurring.

#### 1) Foodborne botulism

The source of the intoxication and other potentially exposed persons must be identified. The case must be interviewed concerning possible food sources. In most cases, this information will need to be obtained from family members or other close contacts, since the case will most likely not be in a condition to be interviewed. Use of the MDPH *Foodborne Illness Complaint Worksheet* (see copy in Appendix A) will facilitate recording more information pertinent to foodborne transmission. Please call the MDPH Division of Food and Drugs at (617) 983-6712 for assistance in determining possible food sources. Use the following guidelines to assist you in the investigation.

- a) Identify all home-canned foods eaten during the week prior to symptoms. The most suspect foods are those eaten less than two days before onset, those that are low in acid and those that were not eaten by other persons that remain well. Keep in mind that some cases may experience symptoms later than the case or that are less severe than those of the case.
- b) Identify all commercially canned foods eaten during the week prior to the onset of illness. For implicated foods, determine the brand, manufacturer, package size, lot number, and place and date of purchase.
- c) Identify all sausage and other preserved meats eaten during the week prior to onset of illness. Meat products that have not been adequately refrigerated should also be suspected.
- d) Identify all smoked or otherwise preserved fish eaten during the week before onset of symptoms.
- e) Identify other potentially exposed persons. Other persons who have eaten implicated food must be reached as soon as possible and advised to seek health care immediately. Depending on the time of ingestion, other exposed persons might be candidates for purging and at the very least should be under close medical supervision.
  - 1. Obtain the name, address, and telephone number of every person who may have eaten the suspected food item.
  - 2. Obtain the name, address, and telephone number of every person who may have the suspect home-processed food in his or her possession.
- f) Remove implicated food items from the environment for testing. The MDPH Division of Food & Drugs (DFD), reachable at (617) 983-6712, will coordinate pickup and testing of food samples. If a commercial product, DFD will also coordinate follow-up with relevant outside agencies.
- 2) Wound botulism: No follow-up required.
- 3) **Infant botulism:** Ask caretakers about honey consumption, otherwise, extensive epidemiological follow-up is not usually required. Education should be provided regarding prevention.
- 4) **Adult infectious botulism:** As with infant botulism, extensive epidemiological follow-up is not usually required. Education should be provided regarding prevention.

4 Botulism January 2001

- e. **Botulism Testing:** In all cases of suspected botulism a determination is made by the Division of Epidemiology and Immunization and the case's healthcare provider, based on available clinical and epidemiological data, about the appropriateness of botulism testing. Arrangements will then be made for the submission of appropriate specimens.
- f. **Botulism Antitoxin:** Antitoxin therapy is only administered to adult patients with foodborne or wound botulism. Antitoxin is a horse serum product and may cause serum sickness in approximately 20% of treated persons. Antitoxin is not indicated in cases of infant botulism. The healthcare provider in consultation with the Division of Epidemiology and Immunization must determine the need for antitoxin therapy. CDC must release and approve its use. If needed, antitoxin will be immediately flown into the nearest airport. Local boards of health should be prepared to assist with logistic arrangements. The decision to administer antitoxin must be made immediately. The longer the wait the less effective it will be. Since testing for the presence of toxin or bacteria in clinical specimens can take many days, the decision to administer antitoxin cannot wait for testing to confirm the infection.
- g. Institution of disease control measures is an integral part of a case investigation. It is the LBOH responsibility to understand, and, if necessary, institute the control guidelines listed below in Section 4), Controlling Further Spread.

## 4) CONTROLLING FURTHER SPREAD

## A. Isolation and Quarantine Requirements (105 CMR 300.200)

**Minimum Period of Isolation of Patient** 

No restrictions.

**Minimum Period of Quarantine of Contacts** 

No restrictions.

#### B. Protection of Contacts of a Case

None.

## C. Managing Special Situations

## Reported Incidence Is Higher than Usual/Outbreak Suspected

Any case botulism is considered an outbreak and must be investigated to determine the source of infection and mode of transmission. See Section 3) C above for the proper response to a case of suspect or confirmed botulism.

*Note:* Refer to the MDPH's *Foodborne Illness Investigation and Control Reference Manual* for comprehensive information in investigating foodborne illness complaints and outbreak. (Copies of this manual were distributed to local boards of health in 1997–98. It can also be located on the MDPH website in PDF format at <a href="http://www.magnet.state.ma.us/dph/fpp/refman.htm">http://www.magnet.state.ma.us/dph/fpp/refman.htm</a>.) For recent changes (fall of 2000) to the Massachusetts Food Code, contact the Division of Food and Drugs, Food Protection Program at (617) 983-6712 or through the MDPH website at <a href="http://www.state.ma.us/dph/fpp/">http://www.state.ma.us/dph/fpp/</a>.

*Note:* If a bioterrorist event is suspected, the MDPH and other response authorities will work closely with local boards of health and provide instructions/information on how to proceed.

#### **D. Preventive Measures**

#### **Personal Preventive Measures/Education**

To avoid future exposures, recommend that individuals:

January 2001 Botulism 5

- Who are interested in home canning and other preservation techniques be educated about the proper time, pressure and temperature required to destroy spores. More information can be obtained from the Massachusetts Division of Food and Drugs.
- Not open bulging containers, and not eat or even "taste-test" foods with off odors.
- Not feed honey to children less than one year old.

## ADDITIONAL INFORMATION

The following is the formal CDC surveillance case definition for botulism. It is provided for your information only and should not affect the investigation or reporting of a case that fulfills the criteria in Section 2) A of this chapter. (CDC case definitions are used by the state health department and CDC to maintain uniform standards for national reporting.) For reporting to the MDPH always use the criteria outlined in Section 2) A.

## Laboratory criteria for diagnosis

*Foodborne*: Detection of botulinum toxin in serum, stool or patient's food or isolation of *C. botulinum* from stool.

*Infant*: Detection of botulinum toxin in stool or serum or isolation of *C. botulinum* from stool.

Wound: Detection of botulinum toxin in serum or isolation of C. botulinum from a wound.

*Other*: Detection of botulinum toxin in a clinical specimen or isolation of *C. botulinum* from a clinical specimen.

#### **Confirmed Case Definition**

*Foodborne:* A clinically compatible case that is laboratory-confirmed or that occurs among persons who ate the same food as persons who have laboratory-confirmed botulism (see laboratory criteria).

*Infant*: A clinically compatible case that is laboratory-confirmed occurring in a child aged < one year.

Wound: A clinically compatible case that is laboratory confirmed in a patient who has no suspected exposure to contaminated food and who has a history of a fresh, contaminated wound during the two weeks before onset of symptoms.

*Other:* A clinically compatible case that is laboratory confirmed in a patient aged  $\geq$  one year who has no history of ingestion of suspect food and has no wounds.

#### REFERENCES

American Academy of Pediatrics. 1997 Red Book: Report of the Committee on Infectious Diseases. Illinois, Academy of Pediatrics, 1997.

CDC. Case Definitions for Infectious Conditions under Public Health Surveillance. MMWR. 1997; 46:RR-10.

CDC Website. Botulism (*Clostridium botulinum*: Frequently Asked Questions). Available at <a href="http://www.cdc.gov/ncidod/dbmd/diseaseinfo/botulism\_g.htm">http://www.cdc.gov/ncidod/dbmd/diseaseinfo/botulism\_g.htm</a>. Updated September 2, 1999.

Chin, J., ed. *Control of Communicable Diseases Manual*, 17<sup>th</sup> Edition. Washington, D.C., American Public Health Association, 2000.

MDPH. *Regulation 105 CMR 300.000: Reportable Diseases and Isolation and Quarantine Requirements.* MDPH, Promulgated November 1998 (Printed July 1999).

Oregon Health Division. Investigative Guidelines: Botulism. Oregon Health Division, November, 1994.

Shapiro, Roger L., *et al.* Botulism in the United States: A Clinical and Epidemiologic Review. *Annals of Internal Medicine*. August 1, 1998; 129:3, pp. 221-228.

6 Botulism January 2001